CONTINUING MEDICAL EDUCATION

The Differential Diagnosis of Hearing Loss

Thomas Zahnert

SUMMARY

Background: According to the World Health Organization, hearing loss is one of the six leading contributors to the global burden of disease. It is becoming an ever more important problem in society at large, not just because the population is aging, but also because young people increasingly spend their leisure time in activities that expose them to excessive noise. On the other hand, the treatment of hearing loss is improving, as the result of technical developments in otological surgery, hearing aids, and cochlear implants. For nearly every type of hearing loss, there is now some type of rehabilitative treatment. The prerequisite to effective care is timely and accurate diagnosis.

Method: Review of the pertinent literature and national guidelines.

Results and Conclusion: The available epidemiological data on hearing loss in Germany are inadequate. It is roughly estimated that 13 to 14 million people in Germany are in need of treatment for hearing loss. The most common types of permanent hearing loss are those associated with old age, chronic otitis media, and acoustic trauma. Transient hearing loss is particularly common in childhood as a result of inadequate ventilation of the middle ear. The further technical development of cochlear implants has now widened their indications to include severe congenital deafness and presbycusis.

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Even mild hearing loss can be a major disadvantage in a world of ever-faster information exchange. People who cannot hear spoken language well enough to process it quickly may find themselves cut off from others at work, at home, or in social situations. New types of rehabilitative treatment are now available for this problem. According to the World Health Organization (WHO), hearing loss is one of the six leading contributors to the burden of disease in industrialized countries: Along with ischemic heart disease, depression, and Alzheimer's disease, it is one of the conditions that most severely impair the quality of life of those who suffer from them (1).

Learning objectives

This article is intended to enable its readers to

- become acquainted with the major types of hearing impairment,
- gain an overview of the classification of hearing impairment that is currently in use, partly for historical reasons, and
- learn about the many available treatments and their potential indications.

Methods

This paper is based on a selective review of literature that was retrieved by a search of the Medline and Cochrane databases (2000–2011), as well as on a guideline concerning peripheral hearing impairment in childhood that was issued by the German Society for Phoniatry and Pediatric Audiology *(Deutsche Gesellschaft für Phoniatrie und Pädaudiologie e. V.)* and on the guidelines on cochlear implants and sudden sensorineural hearing loss issued by the German Society for Otolaryngology and Head and Neck Surgery *(Deutsche Gesellschaft für Hals-Nasen-Ohren-Heilkunde, Kopf- und Hals-Chirurgie)*.

Definition

The term "hearing impairment" refers to a lessening of hearing ability in the widest possible sense, ranging from subjectively barely appreciable impairments to total deafness.

TABLE 1

The WHO classification of the severity of hearing impairment, with general clinical recommendations[*1]

Grades of hearing impairment	Mean hearing loss in pure-tone audiogram	Clinical findings	Recommendations
0 – No impairment	25 dB or better	No or very slight hearing problems. Able to hear whispers.	Counseling, follow-up examination; if conductive hearing loss is present, evaluate indication for surgery
1 – Slight impairment	26–40 dB	Able to hear and repeat words spoken in normal voice at 1 meter.	Counseling, hearing aids may be advisable; if conductive hearing loss or combined hearing loss is present, surgical treatment may be indicated
2 – Moderate impairment	41–60 dB	Able to hear and repeat words spoken in raised voice at 1 meter.	Hearing aids recommended; if conductive hearing loss or combined hearing loss is present, surgical treatment may be indicated
3 – Severe impairment	61–80 dB	Able to hear some words when shouted into better ear.	Hearing aids needed; if an external hearing aid is not possible, consider an implanted hearing aid or cochlear implant; lip-reading and signing for supportive treatment
4 – Profound impairment including deafness	81 dB or higher	Unable to hear and understand even a shouted voice.	Failure of a hearing-aid trial is now usually considered an indication for a cochlear or brainstem implant; lip-reading and signing can be taught in addition

[*1] The mean hearing loss is calculated separately for each ear as the mean value of hearing loss for the four frequencies 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz.
Modified from WHO: Grades of hearing impairment; www.who.int/pbd/deafness/hearing_impairment_grades/en/index.html

Terminology

The term "hearing impairment" (synonyms: "hardness of hearing," "hypacusis") refers to a lessening of hearing ability in the widest possible sense, ranging from subjectively barely appreciable disturbances to total deafness. Hearing impairment is caused by a disturbance of the conduction of sound to the inner ear, the perception of sound by the sensory cells of the cochlea, or the processing of sound in the cochlear nerves, the auditory pathway, or the cortical auditory centers. Thus, hearing impairment is a symptom of many different diseases that affect the organs of hearing. It is distinct from other hearing disturbances such as hyperacusis (oversensitivity to sound), fluctuating hearing, and tinnitus.

The prevalence of hearing impairment in Germany

According to epidemiological studies, the prevalence of hearing impairment that is severe enough to require treatment is about 19% in Germany (2). This figure is arrived at when hearing impairment is operationally defined as a diminution of hearing ability by at least 40 dB in five test frequencies from 0.5 to 4 kHz. Thus, in 2001, there were about 13.2 million persons with hearing impairment living in Germany. The actual number may be even higher, however, because children up to age 14 were not included in the study, and also because the WHO sets a lower threshold (25 dB) for the definition of hearing impairment.

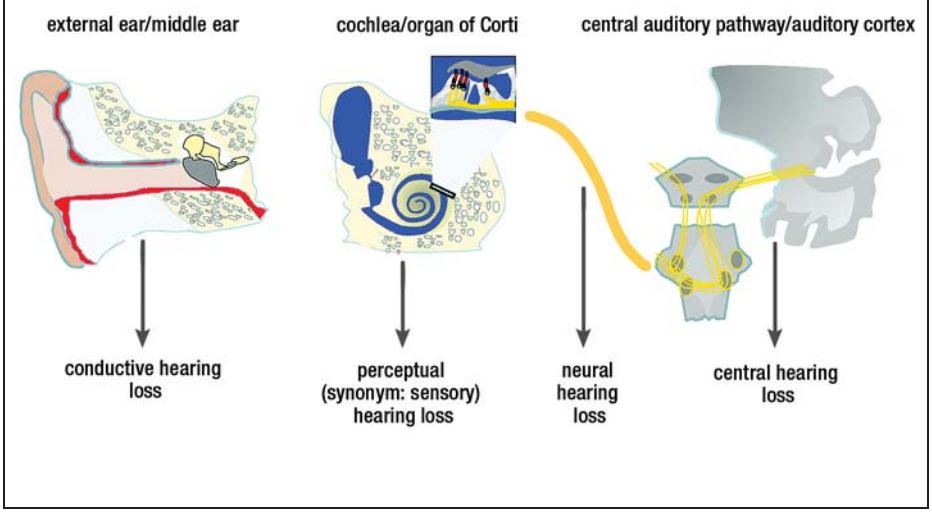
Prevalence

According to epidemiological studies, the prevalence of hearing loss that is severe enough to require treatment is about 19% in Germany.

Congenital bilateral hearing loss

The prevalence of congenital, permanent, bilateral hearing loss is 1.2 per 1000 neonates.

FIGURE 1



Topographic-functional classification of hearing impairment
according to the level of the lesion in the organs of hearing

No study has yet addressed the question of the relative prevalence of the various types of hearing impairment (classified by cause).

The most common type of hearing impairment in childhood is transient conductive hearing loss due to a tympanic effusion. 10% to 30% of children suffer from this problem before their third birthday, with a prevalence as high as 8%. Congenital, permanent, bilateral hearing loss is much rarer, with a prevalence of 1.2 per 1000 children. In adulthood, the most common type of hearing impairment is the sensorineural hearing loss of old age (presbycusis), which affects 40% of all persons aged 65 or older. The next most common types are permanent conductive or combined hearing loss due to chronic otitis media (prevalence 1.5%) and hearing impairment due to acoustic trauma (prevalence 0.05%) (3–8).

Clinical features

Persons with early hearing impairment can often compensate for it for a relatively long time, e.g., by turning up the volume of the radio or television set or (in unilateral hearing impairment) by turning the healthy ear to the sound source. As hearing impairment worsens, vision is used as an additional aid to speech recognition, with an increasing reliance on lip-reading. Common hallmarks of hearing impairment are repeated questioning about things that have not been properly heard, inappropriate answers to misheard questions, and an excessively loud speaking voice.

The state of the evidence for treatments of hearing impairment

Randomized trials have been performed on middle-ear surgery and on the provision of implantable hearing aids and cochlear implants. Poorer evidence is available from clinical trials on the pharmacotherapy of acute inner-ear disorders, in particular sudden sensorineural hearing loss. It can now be said that nearly every kind of permanent hearing loss is treatable.

The classification of hearing impairment

The main classifications that are currently in clinical use are based on the severity of hearing impairment, as assessed by pure-tone audiometry *(Table 1),* and on the basic topographic and functional distinction between conductive hearing loss, sensorineural hearing loss and central hearing loss *(Figure 1).* Other types of classification are by age (e.g., hearing impairment in childhood vs. in old age), temporal course, severity, and the pattern of variation of the auditory threshold as a function of frequency on audiograms (9, 10).

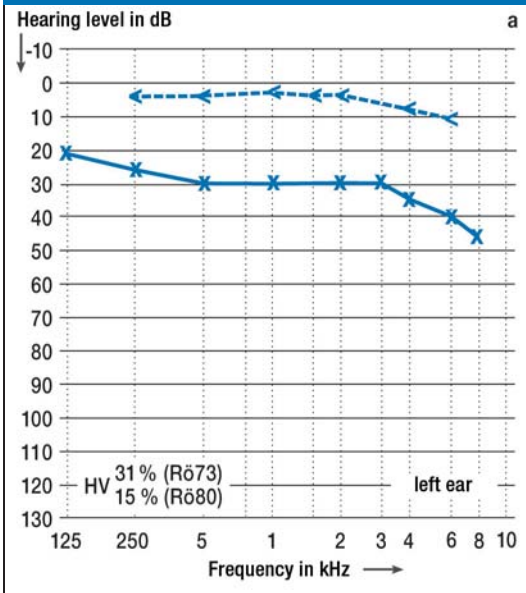
Clinical features

Typical behaviors include turning up the volume of the radio or television set or (in unilateral hearing impairment) turning the healthy ear to the sound source.

The classification of hearing impairment

The basic topographic and functional distinction is between conductive hearing loss, sensorineural hearing loss, and central hearing loss.

FIGURE 2



a) Conductive hearing loss: testing with ear and bone loudspeakers. The cause lies in the external or middle ear. Pure-tone audiometry reveals a difference between the air conduction (x-x-x) and bone conduction (<-<-<) thresholds.

b) Sensorineural hearing loss: testing with ear and bone loudspeakers. The cause lies in the cochlea. Pure-tone audiometry reveals superposable air and bone conduction curves.

Conductive and sensorineural hearing loss

Sound waves are conducted via the external ear and the external auditory canal to the tympanic membrane, which is thereby set in vibration like the membrane of a microphone. These mechanical vibrations are then transmitted by way of the ossicles of the middle ear to the cochlear perilymph and endolymph. All of the disturbances that can arise along the sound conduction pathway are mechanical in nature and are collectively termed conductive hearing loss *(Figure 2)*.

Sound waves can also be conducted to the inner ear through bone. If the skull is acoustically stimulated, e.g., with a tuning fork, the basilar membrane of the cochlea vibrates just as it does when it is set in motion by vibration of the stapes. This pathway of sound conduction, from the skull to the inner ear, is called bone conduction. If sound conducted through bone cannot be heard properly, the disturbance must lie in the cochlea or in more central components of the auditory pathway. Sensory hearing loss, by definition, is hearing impairment due to dysfunction of the cochlea; neural hearing loss is due to dysfunction of the cochlear nerve; and central hearing loss is due to dysfunction of the central auditory pathway or the auditory cortex *(Figures 1 and 2)*. The term "sensorineural hearing loss" refers to hearing impairment due to combined dysfunction of the cochlea and the cochlear nerve.

Common causes of hearing impairment, classified by anatomic site

External auditory canal

Conductive hearing loss: Conductive hearing loss can be due simply to obstruction of the external auditory canal, as by an occluding ceruminal plug or by canal atresia. The latter causes up to 60 dB of hearing loss; if both external auditory canals are atretic in a newborn baby, bone-conduction hearing aids should be provided in the first two to three months of life to enable normal development of hearing and speech.

Tympanic membrane, tympanic cavity, and auditory ossicular chain

Transient conductive hearing loss: Researchers conducting experiments on themselves have shown that closure of the Eustachian tube reduces the pressure in the tympanic cavity to 165 mmWs (daPa) within two hours (11) and thereby lessens the vibration of the tympanic membrane, mainly in the lower frequencies (e1). If the Eustachian tube is blocked for months, a

Tone audiogram

- In sensorineural hearing loss, the air and bone conduction curves in the tone audiogram are superposable.
- In conductive hearing loss, the air and bone conduction thresholds have different curves.

Common types of hearing impairment

- Transient conductive hearing loss in early childhood
- Permanent sensorineural hearing loss in old age

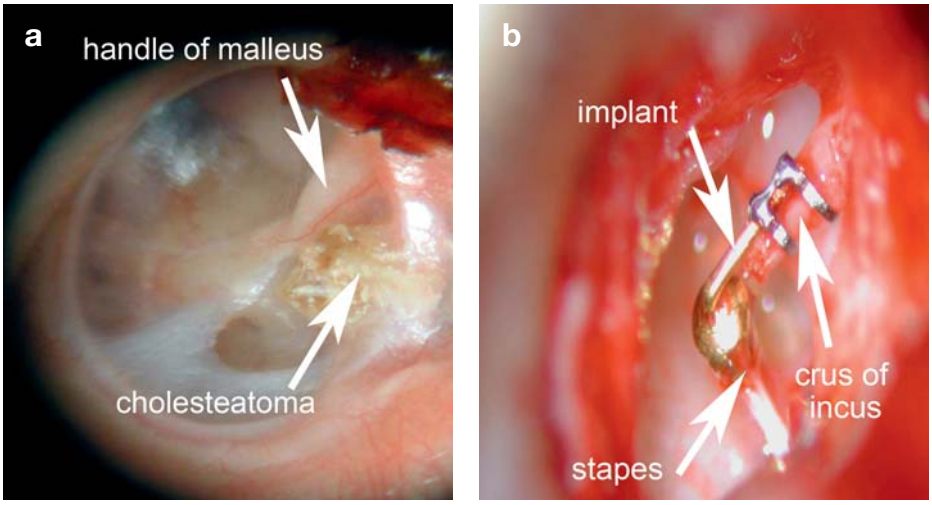


Figure 3:
a) Small cholesteatoma in the upper posterior quadrant of the tympanic membrane with destruction of the crus of the incus
b) View after reconstruction with an angular prosthesis (as described by Plester), bridging the defect between the incus and the stapes

mucoserous tympanic effusion arises, leading to conductive losses of up to 40 dB over the entire frequency range. Tubal blockage is more common in children than in adults because they have narrower Eustachian tubes and are more susceptible to middle ear infection. 10% to 30% of all children have a tympanic effusion leading to conductive hearing loss at some time before their third birthday (3). This problem is much more likely to occur in children with anatomical malformations of the palate and Eustachian tubes, including cleft lip, maxilla, and palate, and in those with Down or Turner syndrome. A tympanic effusion that has been present for three months or longer should be treated with tympanic drainage, as well as adenotomy if necessary, in order to prevent a disturbance of speech development (6).

Middle ear

Permanent conductive hearing loss: Permanent conductive hearing loss is generally caused by chronic bacteria infection of the middle ear, affecting either the mucosa (otitis media mesotympanalis) or the bone (cholesteatoma). Hearing is impaired as a result of muffling of sound by granulations or cholesteatoma as well as enzymatic destruction or inflammatory fixation of the tympanic membrane and ossicular chain. The degree of hearing impairment (in the 30-to-60-dB range) is poorly correlated with the extent of tissue destruction, because inflammatory tissue in the tympanic cavity can itself conduct acoustic vibrations and thus partly compensate for a pathological deficit (12). The treatment of choice is surgery, with the twin goals of eradicating infection and reconstructing the ossicular chain *(Figure 3)*.

Otosclerosis

Otosclerosis, which has a clinical prevalence of 0.3% to 0.4%, consists of fixation of the stapes leading to up to 40 dB of conductive hearing loss in the lower frequencies (13). It is the result of remodeling processes in the bony cochlear wall that can ultimately affect the stapes and the membrane of the round window. Its presumed causes include inflammation (autoimmune processes, measles virus) as well as genetic, metabolic, and hormonal factors (e2). It is most common between the ages of 15 and 40 years and is twice as common in women as in men. Hearing impairment due to otosclerosis can now be treated successfully in 94% of cases with microsurgical stapedoplasty (14).

Perceptual hearing impairment (synonym: sensory hearing loss)

The organ of Corti is the functional unit that transduces perilymphatic vibrations into neural signals *(Figure 4)*. Vibration of the basilar membrane leads to mechanical deflection of the stereocilia and thereby to electrolyte influx into the hair cells, causing depolarization. Motor proteins in the cell walls of the outer hair cells bring

Otosclerosis

The presumed causes of otosclerosis include inflammation (autoimmune processes, measles virus) and genetic, metabolic, and hormonal factors (e2). It is most common from age 15 to age 40 and is twice as common in women as in men.

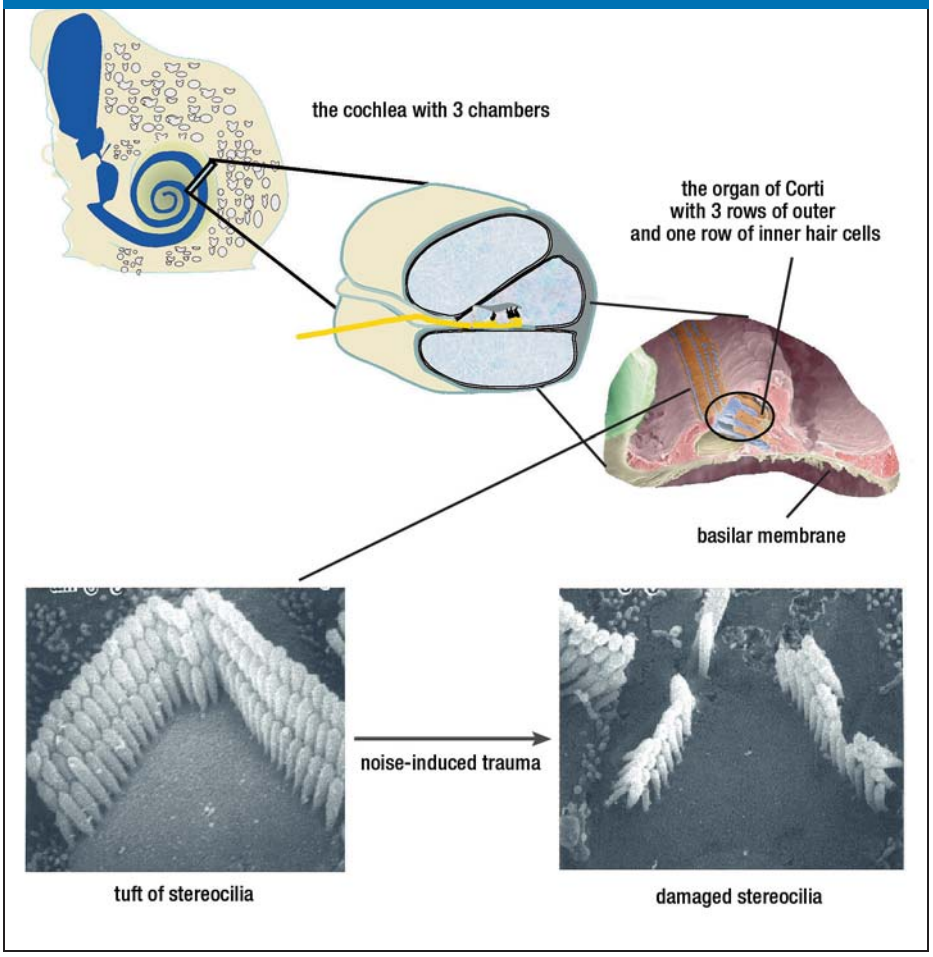
Sensory hearing loss

Sensory hearing loss is due, by definition, to dysfunction of the organ of Corti. The problem is most commonly located in the external hair cells.

The organ of Corti, with outer and inner hair cells (diagram of the human cochlea) and the attached stereocilia before and after noise-induced trauma (chinchilla).

1. Wth the kind permission of Dr. Roger C. Wagner, Professor Emeritus of Biological Sciences, University of Delaware, Newark, Delaware 19716 USA. www.udel.edu/biology/Wags/histopage/histopage.htm
2. Modified from Fukushima N, White P, Harrison R V: Influence of acoustic deprivation on recovery of hair cells after acoustic trauma. Hearing Res. 1990; 50 (1–2): 107–18

FIGURE 4



about a non-linear amplification of this mechanoelectrical transduction process in a particular dynamic range (15). Sensory hearing loss is due, by definition, to dysfunction of the organ of Corti. The problem is most commonly located in the outer hair cells and manifests itself in such cases as a shift of the bone conduction threshold by up to 50 dB, a loss of non-linear reinforcement (the so-called recruitment phenomenon), and impaired frequency selectivity (distortions).

Permanent sensorineural hearing loss in childhood

The prevalence of congenital, bilateral, permanent sensorineural hearing loss of 35 dB or more is estimated at 1.2 per 1000 live births in Germany (16). 25% of these cases can be demonstrated to be of genetic origin, while 18% are acquired, and 57% are of indeterminate cause *(Table 2)*. Congenital hearing loss worsens between the ages of 2 to 5 years in 30% of affected children; thus, even mild hearing loss in

Congenital hearing loss

Congenital hearing loss worsens between the ages of 2 to 5 years in 30% of affected children; thus, even mild hearing impairment in childhood should be re-evaluated every three to six months.

Congenital syndromes causing hearing impairment

More than 300 different congenital syndromes are associated with hearing impairment.

TABLE 2

Synopsis of the causes and clinical features of hearing impairment, with differential diagnoses for each hearing impairment syndrome

	Conductive hearing loss	Sensory hearing loss	Neural hearing loss	Central hearing loss
Cause	– acoustic-mechanical disturbance of sound conduction in the external auditory canal, across the tympanic membrane, or in the ossicular chain	– dysfunction of the hair cells or their synaptic connections to the cochlear nerve; if the outer hair cells are affected, loss of cochlear amplification and thus of recruitment of intermediate intensities – blurring of frequency resolution – reduction of temporal resolution	– cochlear nerve dysfunction – delayed impulse conduction – disturbed neural encoding of the acoustic signal	– dysfunction of the audtiory pathway or auditory cortex (processing of bilateral auditory stimuli, synchronization, signal modulation, recognition, noise suppression)
Clinical features	If the cause is in the external auditory canal: – reduced sound intensity (sound is perceived as soft) If the cause is in the tympanic membrane or ossicular chain: – altered sound frequency and intensity (high and low tones may be either softer or louder)	– loss of intensity and dynamics – soft noises or speech may be perceived as either too soft or too loud – often, distorted perception	– similar to sensory hearing loss, but usually unilateral – speech perception worse than tone perception	– there may be no disturbance of tone perception – impaired rapid speech processing – impairment of sound localization, poor understanding of speech with superimposed noise, impairment of auditory memory
Differential diagnosis	Acute: – blockage by cerumen – tubular catarrh – tympanic effusion – traumatic eardrum perforation – acute otitis media or externa Permanent: – canal stenosis/atresia – defect of eardrum or ossicular chain due to chronic purulent infection of the mucosa – cholesteatoma – malformation – otosclerosis – tympanosclerosis	Acute: – idiopathic sudden sensorineural hearing loss – acute noise-induced trauma – blast trauma – explosion trauma – bacterial/viral labyrinthitis Hereditary/permanent:: – hereditary hearing impairment – presbycusis – noise-induced hearing impairment – toxic (incl, drug-induced) hearing impairment – idiopathic chronic progressive hearing impairment – drug side effects – lasting sequelae of infections and sudden hearing loss	– acoustic neuroma (= vestibular schwannoma) – other tumors of the petrous bone or cerebellopontine angle (meningioma, chordoma, chondrosarcoma) – compression syndrome	– infarction – hemorrhage – tumor – multiple sclerosis – auditory processing disorder
Audiological testing	– tuning-fork test – whispering test – test of hearing at a distance – pure-tone audiogram – impedance audiometry	– tuning-fork test – whispering test – test of hearing at a distance – pure-tone audiogram – speech audiogram – otoacoustic emissions	– pure-tone audiogram – speech audiogram – supraliminal tests – auditory fatigue tests – electric response audiometry	– test of hearing at a distance – pure-tone audiogram – speech audiogram – supraliminal tests – auditory fatigue tests – electric response audiometry

The classification of hearing impairment

- Conductive hearing loss
- Sensory hearing loss
- Neural hearing loss
- Central hearing loss

Components of audiological testing

- Simple clinical tests: tuning fork, hearing at a distance
- Pure-tone and speech audiometry, impedance measurement, supraliminal tests
- Otoacoustic emissions, electrical reaction audiometry

BOX 1

The causes of congenital sensorineural hearing loss and their percentage distribution

- **Unknown causes (57%)**
- **Acquired (18%)**
  - infectious:
    toxoplasmosis, cytomegalovirus, herpes viruses, mumps, measles, meningitis, sepsis
  - metabolic:
    asphyxia, hyperbilirubinemia
  - toxic:
    alcohol, thalidomide, quinine
  - birth trauma:
    (intracranial hemorrhage, skull trauma, noise)
- **Genetic causes (25%)**
  - among which: non-syndromic (70%):
    autosomal recessive (80%)
    autosomal dominant (17%)
    X-chromosomal (3%)
  - among which: syndromic (30%), e.g.*1:
    Alport syndrome (with progressive renal failure)
    Pendred syndrome (with goiter due to faulty iodine metabolism)
    Cogan syndrome (with interstitial keratitis)
    Waardenburg syndrome (with partial albinism and lateral displacement of the lacrimal puncta)
    Usher syndrome (with retinitis pigmentosa)
    Osteogenesis imperfecta (collagen disorder, bony fractures)
    Goldenhar syndrome (malformation of the pinna, dysmorphic facies)
    Pierre-Robin syndrome (micrognenia, cleft palate)
    Franceschetti syndrome (craniofacial dysmorphism)

*1Only a few of the more than 300 congenital hearing loss syndromes are named here by way of illustration. Modified from (4, 6)

childhood should be re-evaluated every three to six months.

Hearing impairment of genetic origin is due to a congenital syndrome in 30% of cases and is non-syndromic in the remaining 70%. Among the non-syndromic cases, the inheritance pattern of hearing impairment is autosomal recessive in 70% to 80%, autosomal dominant in 10% to 25%, and X-linked in 2% to 3% (6). More than 300 different congenital syndromes are associated with hearing impairment; the main ones are listed in *Box 1*. Non-syndromic, autosomal recessive hearing loss (the most common type) is often due to a genetic mutation that impairs the synthesis of the transmembrane proteins connexin 26 and 30, which in turn affects the ion transport mechanism in the hair cells (gap junction protein). Whenever hearing impairment of genetic origin is suspected, connexin 26 and 30 mutations should be sought. Genetic hearing impairment of recessive inheritance is usually severe; it is usually due to a sporadic mutation, and therefore hard to diagnose (17).

Infectious, toxic, and traumatic processes frequently cause acquired hearing impairment in the pre-, peri-, or postnatal period and can also cause progressive or newly acquired hearing loss later on in childhood. Children with known risk factors should undergo thorough auditory testing *(Table 2)*.

In Germany, neonatal auditory screening for the early detection of hearing impairment has been legally mandated since 2008. The goal is to detect hearing impairment of 35 dB or more in the first three months of life so that any required treatment can be initiated before the age of six months. The mandatory diagnostic evaluation involves objective audiometric techniques:

- transitory evoked otoacoustic emissions (TEOAE) and/or
- automatic auditory brainstem response (AABR).

The treatment of hearing impairment in childhood is interdisciplinary, with the early provision of hearing aids and the active promotion of hearing and speech acquisition in pediatric audiology centers. Congenitally deaf or hearing-impaired children whose auditory threshold exceeds 90 dB are better served with cochlear implants than with conventional hearing aids (e3). Speech acquisition is best when bilateral cochlear implants are provided before the child's second birthday (18).

Acute sensory hearing loss in adulthood

Acute hearing impairment resulting from damage to the sensory cells of the inner ear, or from impaired inner ear homeostasis, can be classified according to etiology as

- traumatic (e.g., skull-base fracture).
- toxic,
- infectious, or
- idiopathic.

Blast trauma (e.g., from firecrackers) typically involves brief exposure to a very loud noise (over 140

Childhood development

Infectious, toxic, and traumatic processes frequently cause acquired hearing impairment in the pre-, peri-, or postnatal period and can also cause progressive or newly acquired hearing loss later on in childhood.

Acute hearing loss due to damage of the sensory cells or disordered homeostasis of the inner ear can be

- traumatic,
- toxic/infectious, or
- idiopathic.

dB) in which the rise of pressure takes place over a very short time (<1.5 ms). In contrast, explosion trauma involves a slower rise of pressure (> 2 ms) and leads to rupture of the tympanic membrane. Both of these types of acoustic trauma cause acute hearing loss with tinnitus that usually lasts for several hours. In Germany, 28 to 107 per 100,000 persons sustain a blast trauma each year on New Year's Eve (19). Another type of acoustic trauma is acute noise trauma, which results from exposure to relatively loud noise over a longer period of time (seconds to hours), e.g., at rock concerts. In such situations, hearing impairment is mainly due, not to structural injury of the sensory cells, but rather to a severe metabolic disturbance (oxidative stress), which can be either reversible or irreversible depending on its duration and can manifest itself symptomatically as loss of hearing in the high-frequency range (ca. 4 kHz), and tinnitus in the same range. Potential treatments include the intravenous infusion of rheological agents and cortisone.

Hearing impairment due to acute toxic damage to the inner ear

Medications and bacterial and viral toxins can reach the inner ear by way of the membrane of the round window, the cerebrospinal fluid, or the bloodstream and irreversibly damage the hair cells. Medications that are well known to be ototoxic include the aminoglycosides, cytostatic agents, loop diuretics, salicylates, and quinine. The risk of hair-cell damage from medications can be reduced by careful monitoring of serum concentrations *(Box 2)*.

Bacterial toxins and inflammatory mediators that are called forth by viral infection can also have a toxic effect on the inner ear (labyrinthitis). Toxic labyrinthitis can occur, for example, in influenza otitis, purulent meningitis, and chronic otitis media. Systemic viral infections (mumps, measles, rubella, cytomegalovirus, HIV) reach the labyrinth by way of the bloodstream (e4).

BOX 2

Ototoxic causes of sensory hearing loss (after e4)

- **Medications**
  - aminoglycoside antibiotics (gentamicin, streptomycin, tobramycin, amikacin)
  - cytostatic agents (cisplatin, cyclophosphamide)
  - diuretics (furosemide, ethacrynic acid)
  - other (salicylates, quinine)
- **Industrial substances**
  - heavy metals (mercury, lead, arsenic)
  - solvents (aminobenzenes, nitrobenzenes)
  - other (carbon monoxide, fluorocarbons, organosulfur compounds, carbon tetrachloride)
- **Abused substances**
  - cocaine, heroin, tobacco, alcohol
- **Viral and bacterial toxins**
  - bacterial toxins (pneumococci, staphylococci, streptococci, Haemophilus) in: otitis media, meningitis, scarlet fever, sepsis, syphilis
  - viral infections: mumps, measles, rubella, influenza, HIV, cytomegalovirus
- **Metabolic causes**
  - vitamin B12 deficiency, hyperlipidemia, folic acid deficiency

Sudden sensorineural hearing loss

As its name implies, sudden sensorineural hearing loss is a syndrome involving sudden hearing impairment that is not due solely to a problem in the inner ear. Its causes include (20):

- systemic infection, 12,8% (e.g., meningitis, syphilis, or HIV infection),
- diseases of the ear, 4.7% (e.g., cholesteatoma),
- trauma, 4.2% (e.g., blast trauma, skull-base fracture),
- cardiovascular disease, 2.8%, and
- paraneoplastic involvement of the inner ear, 2.2%.

In 71% of cases, however, the diagnostic algorithm fails to reveal any cause (idiopathic sudden sensorineural hearing loss). Models of the etiology and pathogenesis of this disorder generally proceed from the assumption of an unknown viral, vascular, or immunological cause that leads to a disturbance of homeostasis in the inner ear (e5).

Permanent sensorineural hearing loss in adulthood

Presbycusis

Presbycusis is bilateral sensorineural hearing loss of multifactorial origin, beginning between age 50 and age 60 and characterized by impaired hearing of high frequencies, as can be revealed by pure-tone audiometry. It is thought to be due to physiological aging

Hearing impairment due to acute toxic damage to the inner ear

Medications that are well known to be ototoxic include the aminoglycosides, cytostatic agents, loop diuretics, salicylates, and quinine.

Ototoxic causes of sensory hearing loss

- Medications, industrial chemicals
- Substances of abuse
- Viral and bacterial toxins
- Metabolic abnormalities

processes, particularly as they affect the microvascular blood supply of the hair cells (sensory type), with resulting ischemia, hypoxia, and oxidative stress. Such processes can also affect the ganglion cells (neural type) or the stria vascularis (metabolic type). About 40% of persons over age 65 suffer from presbycusis (7). There is no known way to prevent it; it can be treated symptomatically with the provision of a hearing aid if the auditory threshold shift is 30 dB or more in the speech-frequency range. Severe hearing impairment in old age with loss of speech comprehension can be treated with a cochlear implant.

Occupationally induced hearing impairment

Noise-related hearing loss accounts for 40% of all cases of recognized occupational disease and is thus the most common of all occupational diseases (e6). The main causative factor is the quantity of energy transferred into the inner ear, i.e., the intensity of noise combined with its duration. Hearing impairment generally arises after years of exposure to noise above 85 dB for the entire working day. Damage to the outer hair cells typically begins in the 4 kHz range (revealed as a C5 drop on pure-tone audiometry) (21).

Another type of hearing impairment due to long-term exposure is chronic, toxic occupationally induced hearing impairment. It is caused by years of exposure to substances such as heavy metals, benzenes, and various other carbon compounds. A typical feature of this kind of hearing impairment is a symmetric rise of the auditory threshold *(Box 2)*.

Neural hearing loss

This category includes all types of hearing impairment due to diseases affecting the cochlear nerve (including its synapses). The common causes of neural hearing loss are tumors (meningioma, acoustic neuroma, chordoma, chondrosarcoma) and inflammatory destruction of the petrous bone (cholesterol granuloma, cholesteatoma).

Acoustic neuroma (more properly, "vestibular schwannoma") is the most common cause of neural hearing loss, with an incidence of 1.74 per 100 000 persons per year (e7). Any patient with unilateral loss of bone conduction on pure-tone audiometry, without any immediately recognizable cause, should undergo brainstem potential recording (BERA) and an MRI scan with contrast medium. The management of acoustic neuroma is now an interdisciplinary matter, as these tumors can be treated either by surgical removal or by stereotactic radiation procedures ("radiosurgery"); for some patients, close observation is a further option (including serial MRI scans and tests of hearing and vestibular function).

Auditory synaptopathy/neuropathy is a special type of neural hearing loss affecting the synapses of the inner hair cells that can arise either in isolation or as a component of a generalized neuropathy. There is a disturbance of temporal processing that may lead to total blockage of impulse conduction (22).

Central hearing loss

The auditory signal, originally consisting of mechanical pressure waves, is processed and encoded in various forms at multiple levels of the auditory pathway. The higher the neural dysfunction is located, the more complex the hearing disturbance; thus, the patient may have difficulty in recognizing certain signals amid acoustic noise, in disentangling simultaneous speech signals, or in recognizing timbre.

Adults can suffer from central hearing impairment as a result of trauma, inflammatory/infectious processes, infarction, and space-occupying lesions (tumors, hemorrhage) that affect the auditory pathway and the auditory centers. Bulbopontine hearing impairment is usually associated with central dizziness, ataxia, and other neurological abnormalities pointing to brainstem involvement. Its diagnosis is confirmed by brainstem audiometry with measurement of the intermediate potentials. Midbrain hearing impairment (e.g., in multiple sclerosis) is accompanied by clinically evident neurological abnormalities (motor and sensory deficits); specific questioning is required to elicit the history of paracusis (false acoustic perception) and diplacusis (hearing of double tones) (23). Special hearing tests can also reveal abnormalities of directional hearing and of rapid speech recognition.

A special type of central hearing loss affecting school-age children is known as "central auditory processing disorder" (CAPD). This is a dysfunction of central, modality-specific auditory processing. It is debated whether CAPD truly constitutes an independent disease entity, as some claim that it is seen only as a component finding in the setting of developmental disorders, intellectual impairment, multimodal perceptual disorders, and activity and attention disorders (24). Its treatment is interdisciplinary and involves specific training of the deficient auditory functions.

Occupationally induced hearing loss

Noise-related hearing loss accounts for 40% of all cases of occupational disease and is thus the most common of all occupational diseases.

Neural hearing loss

This category includes all types of hearing impairment due to diseases affecting the cochlear nerve (including its synapses). The common causes of neural hearing loss are tumors and inflammatory destruction of the petrous bone.

Conflict of interest statement
Prof. Zahnert has received reimbursement of travel fees for lectures and workshops as well as scientific lecture honoraria from the following companies: Med-El Deutschland, Cochlear GmbH, ATOS, MIP Pharma GmbH, Merck Pharma GmbH, and HNO-Update GmbH. He is active as a consultant and is engaged in scientific collaboration with the following companies, with financial support of research projects: Kurz-Medizintechnik GmbH, Med-El, Cochlear GmbH, and Omega Consulting GmbH. Prof. Zahnert has 11 patents pending for middle-ear implants and implantable hearing systems.

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The solutions to the following questions will be published in issue 33/2011. The CME unit "Current Pacemaker and Defibrillator Therapy" (issue 21/2011) can be accessed until 8 July 2011.

For issue 28–29/2011, we plan to offer the topic "Hormonal Contraception."

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Please answer the following questions to participate in our certified Continuing Medical Education program. Only one answer is possible per question. Please select the answer that is most appropriate.

Question 1

Defects of the tympanic membrane cause what kind of hearing impairment?

a) Conductive hearing loss
b) Perceptual hearing loss
c) Sensory hearing loss
d) Central hearing loss
e) Auditory processing disorder

Question 2

What is the most common cause of transient hearing impairment in childhood?

a) Acute purulent otitis media
b) Congenital malformation of the middle ear
c) Otosclerosis
d) Tympanic effusion
e) Chronic otits media

Question 3

Which of the following classes of medications is not oto-toxic?

a) Loop diuretics
b) Cytostatic agents
c) Salicylates
d) Corticosteroids
e) Aminoglycosides

Question 4

What is most commonly found to be the cause of sudden sensorineural hearing loss?

a) A local disease of the ear
b) A viral infection
c) A cardiovascular disease
d) An acoustic neuroma
e) No determinable cause

Question 5

What frequency is most severely affected in incipient noise-induced hearing impairment?

a) 500 Hz
b) 1 kHz
c) 8 kHz
d) 4 kHz
e) 15 kHz

Question 6

From what age onward is there a 40% prevalence of presbycusis?

a) From age 25 onward
b) From age 35 onward
c) From age 45 onward
d) From age 55 onward
e) From age 65 onward

Question 7

Which of the following diagnoses is associated with central hearing loss?

a) Menière's disease
b) Otosclerosis
c) Auditory processing disorder
d) Acoustic neuroma
e) Presbycusis

Question 8

A patient complains of difficulty hearing, stating that he cannot understand or carry on a conversation from a distance of more than 1 meter. His pure-tone audiogram shows an intermediate degree of hearing loss (30 dB).

According to the WHO classification, how severe is this man's hearing impairment?

a) Grade 0, no impairment
b) Grade 1, slight impairment
c) Grade 2, moderate impairment
d) Grade 3, severe impairment
e) Grade 4, profound impairment including deafness

Question 9

During what period of time should neonatal auditory screening for the detection of congenital hearing loss be performed?

a) When the baby is no more than 30 days old
b) When the baby is no more than 3 months old
c) When the baby is no more than 7 days old
d) On the day the baby is born
e) When the baby is no more than 2 weeks old

Question 10

During what period of time does congenital hearing loss tend to progress?

a) In the first six months
b) In the second half of the first year
c) Before the third birthday
d) From the first to the third birthday
e) From age 2 to age 5

CONTINUING MEDICAL EDUCATION

The Differential Diagnosis of Hearing Loss

by Thomas Zahnert

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